2772-9931
https://doi.org/10.1016/j.atssr.2023.02.020

Lung: Case Report

Ventral Scapular Osteochondroma in Hereditary Multiple Exostosis

Gregory P. Mouradian, MD,[1,2]
Sam Hong, MD,[1,3] and
Emmanuel Daon, MD[1,3]

Hereditary multiple exostosis is a rare, autosomal dominant, highly penetrant genetic disorder characterized by the development of osteochondromas on the metaphysis of long and flat bones. The development of osteochondromas on the scapula is unusual but has been reported. Here, we present a case of an osteochondroma of the ventral scapula with preoperative concern for invasion into the second rib. Through an open approach, the lesion was found to be well isolated within a bursa, and it was resected without complication. The patient had a short hospital stay with good functional and cosmetic results.

(Ann Thorac Surg Short Reports 2023;1:246-248)

Hereditary multiple exostosis (HME), also known as hereditary multiple osteochondromas, is a rare genetic disorder that affects 1 of 50,000 people in the United States. It is inherited in an autosomal dominant fashion through the genes encoding exostosin glycosyltransferase 1 (*EXT1*) and exostosin glycosyltransferase 2 (*EXT2*) with nearly 100% penetrance.[1] Patients with HME have osteochondromas at the metaphysis of long and flat bones that are appreciated clinically in early childhood, most frequently around the knee. Radiographs are used to identify symptomatic or asymptomatic growths.

Lesions from flat bones like the pelvis and scapula are rare but reported in the literature. These osteochondromas carry a risk of malignant degradation of about 2% to 5%, which increases with age. Patients with scapular lesions usually complain of pain with range of motion and popping or locking sensation of the shoulder. Crepitus is frequently appreciated. Whereas exostoses of the shoulders are common within patient cohorts, occurring in about 85% of patients, exostoses of the scapula are rare, occurring in only 3% of all lesions. Those with scapular involvement tend to have a heavier burden of exostoses and possess the *EXT1* mutation. Scapular exostoses have a higher propensity for malignant transformation.[2] In rare cases, large thoracic osteochondromas have been reported to erode into the pleural space, resulting in hemothorax[3] or pneumothorax.[4] Lesions have been reported to invade the diaphragm, causing ruptures,[5] and the mediastinum, compressing the coronary arteries.[6] Despite documentation of their known incidence, there is a paucity of literature on ventral scapular osteochondromas, making surgical management of these lesions complex.

Our patient is a 26-year-old right-handed man with a known history of HME diagnosed in childhood. Before evaluation, he had undergone multiple bone excisions of previous osteochondromas on his bilateral fibulas and femurs. He described painful popping and locking of his right shoulder that prevented activities requiring full range of motion. Previous interventions and resections for osteochondromas were uneventful, and he recovered appropriately. Previous pathologic examination revealed benign histologic type. He did not have diabetes or use tobacco products.

A computed tomography scan was obtained of the right upper extremity and chest. It demonstrated a large osteochondroma projecting anteriorly and inferiorly from the superior scapular border to within millimeters of the right posterolateral second rib (Figure 1). With the possible need for rib resection, a multidisciplinary approach was used with a thoracic and orthopedic surgery combination case.

The patient was placed in a left lateral decubitus position with appropriate axillary support and bone protection. We made a limited, high, and right posterior thoracotomy incision along the right scapula. The trapezius and rhomboid muscles were divided sharply through to the tissue plane between the posterior chest wall and scapula. In this space, we

Accepted for publication Feb 27, 2023.

[1]The University of Kansas Medical Center, Kansas City, Kansas; [2]Department of Surgery, University of Kansas School of Medicine, Kansas City, Kansas; and [3]Division of Cardiothoracic Surgery, University of Kansas School of Medicine, Kansas City, Kansas

Address correspondence to Dr Mouradian, Department of Surgery, University of Kansas School of Medicine, 3901 Rainbow Blvd, Mail Stop 2005, Kansas City, KS 66160; email: gmouradian@kumc.edu.

encountered a large bursa that had isolated the osteochondroma from surrounding structures. The osteochondroma was easily palpated on the ventral surface of the scapula (Figure 2). The tumor was resected with an osteotome and rongeurs until the ventral surface was smooth.

The tumor did not invade the second rib or any other area of the chest wall. The pleural space was not entered. In the operating room, we tested and achieved full passive range of motion with the right arm and shoulder without limitations or hinge points. Hemostasis was achieved. The rhomboid and trapezius muscles were reapproximated. The skin was closed.

The patient was managed postoperatively with a sling and instructed to not bear weight with the right upper extremity. He was discharged home on postoperative day 1. He was seen in clinic about 2 weeks later and found to be progressing appropriately. His pain had resolved completely, and he had a full range of motion in his right shoulder. Pathologic examination was consistent with an osteochondroma, without evidence of malignant transformation.

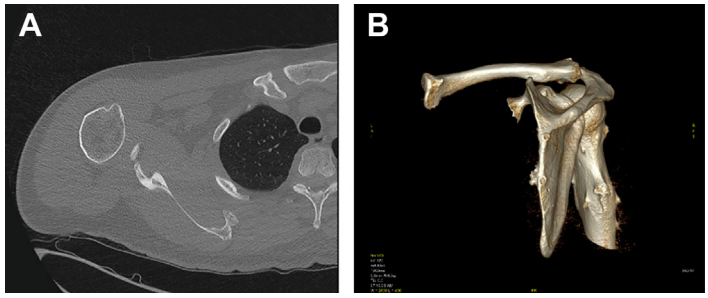

FIGURE 1 (A) Preoperative computed tomography scan. (B) Three-dimensional reconstruction of the right shoulder with a ventral osteochondroma.

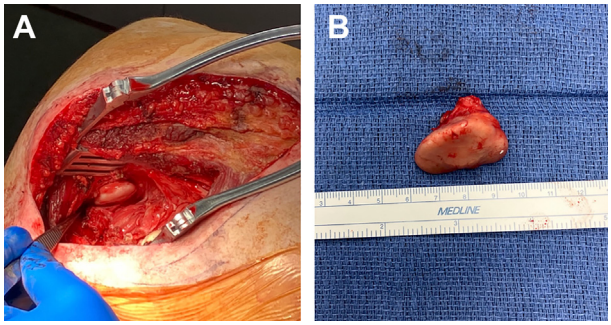

FIGURE 2 (A) Right scapula retracted up divided trapezius and rhomboid with tumor in situ. (B) Osteochondroma, measuring about 3.5 cm, excised.

COMMENT

HME is a rare autosomal dominant genetic disorder characterized by the development of osteochondromas, predominantly on long bones. With a slight male predominance, HME is mainly driven by *EXT1* and *EXT2* gene mutations, often tracked within known family lineage. The development of osteochondromas on the thorax is rare and can be accompanied by pulmonary complications in advanced disease as a result of mass effect. In particular, osteochondromas of the scapula are unusual, and growths on the ventral surface are rarer still. Patients with scapular osteochondromas present with painful shoulder movement as well as with clicking or crepitus. Plain films and computed tomography scans are essential for diagnosis and delineation of anatomy. Although these bone growths have low malignant potential—about 2%—some data suggest that growths on the scapula have a greater probability for malignant transformation. Surgical intervention is indicated when symptoms are poorly controlled or there are concerns for malignant transformation. Here we describe the case of a 26-year-old man with history of HME presenting with a ventral scapular mass. Preoperative imaging was concerning for involvement of the second rib. Resection was performed through a dorsal approach by dividing the trapezius and rhomboid. The osteochondroma had a surrounding bursa that isolated the mass from the second rib. After resection, the patient had an uneventful recovery with good functional outcome and cosmesis.

FUNDING SOURCES
The authors have no funding sources to disclose.

DISCLOSURES
The authors have no conflicts of interest to disclose.

PATIENT CONSENT
Obtained.

Ann Thorac Surg Short Reports
2023;1:246-248

REFERENCES

1. Wuyts W, Schmale GA, Chansky HA, et al. Hereditary multiple osteochondromas. In: GeneReviews [Internet]. University of Washington, Seattle; August 3, 2000 [updated August 6, 2020].

2. Clement ND, Ng CE, Porter DE. Shoulder exostoses in hereditary multiple exostoses: probability of surgery and malignant change. *J Shoulder Elbow Surg*. 2011;20:290-294. https://doi.org/10.1016/j.jse.2010.07.020

3. Lin CY, Chang CC, Chuang MT. Spontaneous haemothorax secondary to rib exostosis. *Heart Lung Circ*. 2017;26:e62-e63. https://doi.org/10.1016/j.hlc.2017.02.005

4. Dumazet A, Launois C, Dury S, et al. Hereditary multiple exostoses of the ribs as an uncommon cause of pneumothorax: a case report. *Medicine (Baltimore)*. 2018;97:e11894.

5. Abdullah F, Kanard R, Femino D, et al. Osteochondroma causing diaphragmatic rupture and bowel obstruction in a 14-year-old boy. *Pediatr Surg Int*. 2006;22:401-403.

6. Rodrigues JC, Mathias HC, Lyen SM, et al. A novel cause of acute coronary syndrome due to dynamic extrinsic coronary artery compression by a rib exostosis: multimodality imaging diagnosis. *Can J Cardiol*. 2015;31:1303.e9-1303.e11.